Editorial

Cerebellar tonsil position and Chiari malformation

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The paper by Dr. Smith and colleagues describes the position and shape of the tonsils in a large group of patients who had undergone MRI of the head or cervical spine for reasons unrelated to abnormalities in the posterior fossa. The authors examined 2400 MR images, having identified 300 scans for patients in each decade of life from the 1st to the 7th and older. A number of important findings are noted. First, the distribution of the location of the tips of the tonsils relative to the foramen magnum is normal in all age decades, except the 1st decade. The distribution in the 1st decade is normal, except for a tail of patients in whom the tonsil tips extend more than 6 mm below the foramen. Second, the average position of the tonsils relative to the foramen magnum increases in each subsequent decade, and the rise is relatively linear from the 1st to the 7th decade. This means that the lowest position of the tonsils that can be considered normal is different for each decade. If the 95th percentile is used as the cutoff for normal, the lowest position of normal tonsils is 7.5 mm below the foramen magnum for ages 0–10 years, 4 mm for 11–20 years, 5 mm for 21–30 years, 4 mm for 31–40 years, 4 mm for 41–50 years, 3 mm for 51–60 years, 2.5 mm for 61–70 years, and 2 mm for 71 years and older. Of note, because of the tail of very low tonsils in the 1st decade, the 95th percentile is skewed to a lower level. If one removes the tail and looks at the normal distribution, the 95th percentile is closer to 4 mm. Interestingly, the average position of the tonsils is lower in females than in males. This difference is most marked in the 1st decade of life, with the 95th percentile cutoff being 6 mm in males and 10 mm in females. The relative rise of the tonsils relative to the foramen magnum by decade of age means that fewer patients in each subsequent decade have tonsils more than 5 mm below the foramen.

While the data presented in this paper are now the best we have for defining the “normal” position of the tip of the tonsils relative to the foramen magnum in each of the specified decades of life, the relationship between tonsil position and symptoms of compressed neural tissue at the foramen magnum is less clear. For example, the authors examined the relationship between the shape of the tonsils and the position of the tip of the tonsils. They describe “pegged” tonsils as those that are pointed rather than rounded, suggesting the tonsils are being compressed into the shape of the dorsal subarachnoid space at the cervicomedullary junction. Among the 2400 scans examined, only 20 patients exhibited tonsils with pegged morphology. Interestingly, 3 (15%) of these 20 had tonsils that were less than 5 mm below the foramen magnum. Similarly, of the 74 patients with tonsils 5 mm or more below the level of the foramen, more than half (54%) had rounded, not pegged, morphology. Of the 2400 patients, 22 carried the diagnosis of symptomatic Chiari malformation Type I (CM-I). Unfortunately, the authors do not indicate the level of the tips of the tonsils in this group, nor do they comment on the relationship between the finding of pegged tonsils and patient symptoms.

The authors do a nice job of cataloging the limitations of their study. Most importantly, the patients did have an MR image that included the region of the foramen magnum for some reason. Thus, they are not truly randomly selected, normal patients. Nonetheless, the authors have tried to mitigate bias by eliminating patients with pathology that might influence tonsil position. This study is clearly the best published to date regarding the distribution of the position of the tonsils on MRI and sets a benchmark for clinical use and future studies.

Given their results, the authors state, “Unfortunately, the current clinical tendency appears to be unreservedly admitting such patients [that is, those with tonsils 5 mm or more below the level of the foramen magnum] to a pathoanatomical group we call CM-I based on tonsil position alone. In view of our results, we believe that this should be reconsidered.” I believe the authors are justified in making this statement and that tonsil position alone does not provide sufficient information regarding the question of whether a patient’s symptoms are caused by compression of neural tissue at the level of the foramen. Clearly, some patients with tonsils more than 5 mm below the foramen magnum are asymptomatic, and some patients with tonsils less than 5 mm below the foramen are symptomatic.

Exactly what criteria should be used for deciding if a given patient’s complaints are related to compression of neural tissue at the foramen magnum is unclear. The authors suggest that additional work relating volume of the posterior fossa to patient symptoms is needed. Similarly, the usefulness of MRI CSF flow studies remains...
undefined. The group at the Children’s Hospital of Alabama, led by Dr. W. J. Oakes, has published its algorithm (based on 500 cases) for decision making regarding the treatment of patients with symptoms referable to CM-I. I suggest this is a good place for the concerned clinician to start. (http://thejns.org/doi/abs/10.3171/2013.3.JNS1339)

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The author reports no conflict of interest.

References

Response
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I thank Dr. Raffel for his insightful comments. He has chosen to highlight the issue of CM diagnosis as it relates to our data. It is my hope that the information presented in our report will lead clinicians to question the value of the traditional imaging definition of CM, which has been based to a large extent on measurements of tonsil position. The distribution of cerebellar tonsil position in the population leads us to believe that arbitrary determinations of abnormality at a particular cutoff point are unhelpful or even misleading. Most neurosurgeons have evaluated a large number of seemingly normal individuals in whom a diagnosis of CM was made on the basis of imaging alone because their tonsil position was more than 5 mm below the foramen magnum. Hopefully, better information on the normal distribution of this metric in the population will allow clinicians to place the imaging results of individual patients into proper context.

Although we have provided percentile data for tonsil position by age, I hope that readers will not use an arbitrary percentile, such as the 95th or even the 99th, as a new definition of abnormal. Such an approach will merely substitute one flawed definition of CM for another. Given currently available information, we think it is best to avoid using tonsil position, whether defined by measurements or percentiles, as the only basis for making a determination of abnormality. Such an approach has the potential to lead to an additional burden on some individuals in the form of unnecessary medicalization and treatment as well as potentially ignoring other individuals with a Chiari-like presentation who do not have a low enough tonsil position to meet such a definition.

Finally, we do not attempt to provide clinical guidance for determining good candidates for Chiari decompression in this article because our data do not support any particular stance on this issue. Nevertheless, we hope that clinical decisions will be influenced by the knowledge that tonsil position varies within the population and that low tonsil position is neither rare nor invariably pathological. We agree with Dr. Raffel that reliable clinical symptoms and examination findings should be given much more weight than imaging results when making treatment decisions for this group of patients.

References

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